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Case report

Papillary cystic neoplasm of the pancreas

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Background

Papillary cystic neoplasm of the pancreas is a rare disorder that occurs most commonly in young women. It has a low potential for malignancy, and the prognosis following resection is favourable.

Case outline

An 18-year-old white girl presented with a palpable mass in the right hypochondrium on physical examination associated with epigastric pain, nausea and vomiting, but no fever. Upper gastro-intestinal endoscopy revealed extrinsic compression of the posterior wall of the antrum and duodenal bulb with no mucosal lesion. Computed tomography (CT) scan and then laparotomy revealed a large tumour adjacent to the hepatic hilum and originating from the head of pancreas. Pancreatoduodenectomy was performed, and a diagnosis of papillary cystic neoplasm of the pancreas was made. There was no evidence of recurrence after 6 years of follow-up.

Discussion

A radical surgical approach is justified for papillary cystic neoplasm of the pancreas because of its biological behaviour, local aggressiveness and low incidence of metastases.

Keywords

papillary cystic neoplasm, pancreas, pancreatic tumour.

Introduction

Papillary cystic neoplasm of the pancreas (PCNP) is a rare disorder that primarily affects young women [1–3]. It is important to differentiate PCNP from other more aggressive tumours because of its low potential for malignancy and favourable prognosis after resection [2,3].

The first cases of PCNP were described by Frantz in 1959 [1,4,5]. This tumour was later characterised by Boor and Swanson, who employed the term 'papillary cystic neoplasm of the pancreas' [1]. Since then, close to 100 additional cases have been reported with a variety of names, including solid papillary epithelial neoplasm of the pancreas, solid-cystic tumour of the pancreas, papillary cystic tumour of the pancreas, papillary epithelial neoplasm of the pancreas and low-grade papillary pancreatic neoplasm [6,7].

We describe the case of a young woman with PCNP who received a radical curative resection (Whipple's operation) in the absence of local or distant metastases.

Case report

An 18-year-old white girl was referred with epigastric pain, nausea and vomiting. Physical examination revealed a palpable mass in the right hypochondrium. There was no fever and no other positive features. Full blood count, biochemical screen and chest X-ray were normal, as were hydatid serology and α-fetoprotein (<5 ng/ml). Upper gastro-intestinal endoscopy revealed an extrinsic compression over the posterior wall of the antrum and duodenal bulb, with no evi-

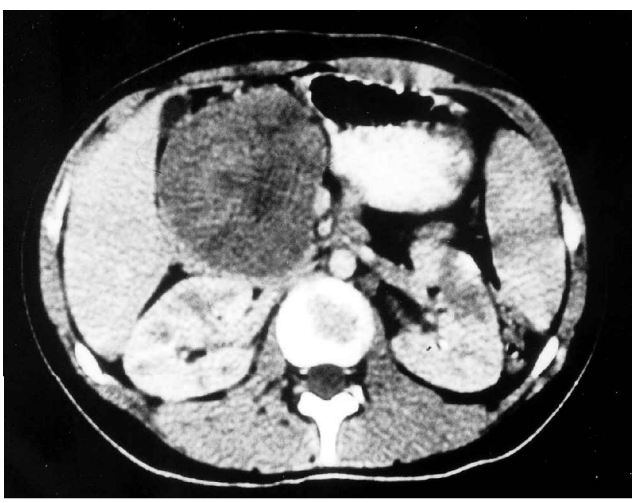

Figure 1. *CT scan showing a mass occupying the region of the hepatic hilum, extending towards the head of pancreas and appearing to involve it.*

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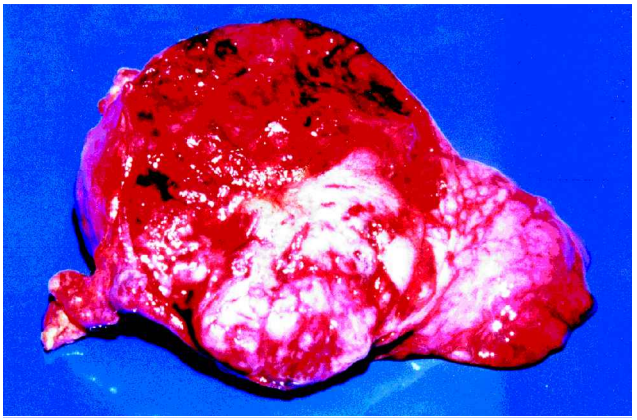

Figure 2. *Resection specimen showing the head and neck of pancreas containing tumour.*

dence of a mucosal lesion. Abdominal ultrasonography and computed tomography (CT) (Figure 1) showed a large tumour, which was thought to originate from the hepatic hilum. Thoracic and cranial CT were normal.

At operation there was a large tumour measuring 8×13 cm in diameter lying close to the hepatic hilum but originating from the head of pancreas. After full dissection to ensure that the portal vein was not involved, a pancreatoduodenectomy (Whipple's operation) was performed (Figure 2). The patient recovered well, and check CT scan at 7 days was satisfactory. She went home 30 days after operation. Up to the present moment, after 6 years of follow-up, the patient has presented no evidence of local recurrence or metastases. She gave birth to a healthy child 5 years and 10 months postoperatively.

The resected specimen comprised the distal stomach, duodenum and head and neck of pancreas, which contained an encapsulated mass. Macroscopic examination showed areas of degeneration, haemorrhagic necrosis, dystrophic calcification and cyst formation. Microscopically, the lesion consisted of small cells with a dark and prominent nucleus and slightly eosinophilic cytoplasm, as well as other papillary formations with fibrovascular cores surrounded by similar cells. There was some necrosis of the tumour cells. The histopathological diagnosis was papillary cystic neoplasm of the pancreas.

Discussion

Papillary cystic neoplasm of the pancreas is a rare condition. Cubilla and Fitzgerald [8] found only one PCNP among 508 cases of epithelial tumour of the exocrine pancreas (0.19%). In addition to the collected series of Compagno *et al.* [9] and Pezzi *et al.* [10], smaller series confirm that PCNP presents as a large tumour with either no symptoms or mild abdominal discomfort and a sensation of postprandial fullness [6,11]. Patients can also present either as an abdominal emergency with haemoperitoneum secondary to post-traumatic or spontaneous rupture [12] or as an incidental finding at laparotomy [1]. Young women are affected in ≥90% of the reported cases [1]; most are non-Caucasian. Mean age at diagnosis is 25 years. The tumour preferentially affects the tail and body of the pancreas. It has the potential for local invasion [5,6,12], but is seldom metastatic [6,8].

According to the literature review carried out by Wang [14], 338 patients with PCNP had been identified up to 1998; in 35% of the cases, the head of pancreas was affected, and in 65% the body and tail (25% and 40%, respectively). Treatment was reported for 44 patients: 13 had been submitted to pancreatoduodenectomy (20%), 21 had their tumours locally resected (32.3%) and 31 were submitted to distal pancreatectomy (47.7%). Survival rate 5 years after diagnosis was 99.12%.

Several other series of PCNP have given results similar to those of Wang. As regards tumour location, in 64% of cases the body and tail of the pancreas were affected, while in 36%, the tumour was located in the head of pancreas. However, results were different in terms of treatment: 24 distal pancreatectomies (61.5%), one total pancreatectomy (2.5%), 11 pancreatoduodenectomies (28.2%) and four local resections (10.2%) were performed. One patient submitted to local resection had to undergo pancreatoduodenectomy 10 years later due to local recurrence and hepatic metastases [1–17].

The case presented here is similar to those described in the literature in the sense that the patient presented with epigastric pain, nausea and vomiting. She was also young and the site affected was the head of pancreas. Despite her age (18 years), we decided to perform a radical procedure (Whipple's operation), with a successful outcome. Another interesting aspect of this case is that whereas most of the patients affected are non-Caucasian, our patient was white.

Accurate diagnosis at the original operation is infrequent due to the rarity of this tumour and its unusual presentation. Preoperatively, the most useful diagnostic

tool seems to be CT scan, which shows a pancreatic mass that is usually well-circumscribed, with solid and cystic components as well as calcifications. Ultrasonography is less efficient for the delineation of such masses, especially if they are calcified or cystic, in which case it cannot show the internal structure. Ultrasonography is valuable for screening and CT for diagnosis [10,13]. A preoperative diagnosis does help to plan appropriate therapy, but definitive diagnosis depends on histological findings. Only a few fine-needle biopsies of this tumour have been reported [2,7,13]. Percutaneous biopsy is more useful for tumours originating from the head of pancreas, whereas excision biopsy is more indicated for those in the body and tail. Cytology is only useful when positive, and its reliability in PCNP is questionable because of variations in the cyst layers and the frequent coexistence of benign and malignant epithelium.

The histogenesis of PCNP is unknown, but both an acinar [12] and a ductal [1] origin have been proposed; endocrine tissue is occasionally found. Origin from a primitive cell capable of endocrine and acinar differentiation may be the likeliest explanation [15]. The predilection for young women suggests a hormonal influence [6,15], while the absence of pancreatic polypeptide hormone on immunohistochemistry militates against a possible origin from islet cells [15,16]. PCNP should not be confused with acinar cell carcinoma of the pancreas, which occurs mainly in adults and affects both sexes equally [13]. It must also be differentiated from mucinous cystic neoplasia, microcystic adenoma (serous cystadenoma) and islet cell tumours, each of which has its own pathological characteristics and prognosis [16].

Although we believe that radical surgery is indicated in all cases, there is evidence that PCNP behaves as a lower-grade malignancy neoplasia in children, thus allowing treatment with local resection when tumours are located in the head of pancreas: radical surgery would then be used in cases presenting recurrence or metastases [17]. In the literature review carried out by Wang [14], more patients below the age of 20 years were treated with local excision (31%) than older patients (15%). The study by Horisawa *et al.* [17], which analysed metastatic cases of PCNP, showed that advanced age on diagnosis or on recurrence increases the possibility of a malignant behaviour. Also, the high recurrence rate in patients with locally resected tumours and the low postoperative morbidity and mortality rates in pancreatic resections carried out by experienced surgeons strongly suggest that a radical and curative resection of the tumour should be performed, regardless of the patient's age [14]. These aspects support our decision to perform a radical surgical procedure (pancreatoduodenectomy) in an 18-year old patient. This decision is also supported by the absence of local recurrence or metastases after 70 months of follow-up.

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